Spinal arteriovenous shunts (AVS) can be separated according to their location. Several entities can thus be recognized:

- Intradural: spinal cord arteriovenous malformations (SCAVM), perimedullary fistulae, radicular AVMs, and filum terminale AVMs;
- Epidural (including paraspinal): epidural AVMs, paraspinous AVMs (PSAVMs), vertebro-vertebral fistula and maxillary arteriovenous fistulae (AVFs) [11].

These lead to neurological symptoms through various pathophysiological mechanisms, including venous congestion, venous compression, and vascular rupture. Hemorrhagic venous infarcts are not seen in the spinal cord.

Intradural AVS are rare, representing only one tenth of central nervous system AVMs in all age groups in the Caucasian population [3]. Very few of these AVS are present in children, although pediatric onset is frequently noted in lesions diagnosed in adulthood. With better imaging technology and higher indices of suspicion, it is now feasible to pick up these lesions early in life. Most are cervical or thoraco-lumbar spinal cord lesions.

Spinal AVS can be classified in two ways, i.e., (1) according to their precise location with respect to the spinal cord, dural, and vertebral elements; and (2) according to potential relationships with genetics, the influence of triggers on the vascular biology, and angiogenesis, in other words as disease entities.
43.2.1
Groups by Location

43.2.1.1
Intracranial Arteriovenous Shunts

These can affect the cord, the nerve roots, or the filum terminale. According to their location, the radiculo-medullary and/or radiculo-pial arteries will feed the spinal cord AVS. They can be found at the surface of the cord or embedded within it, and drain through spinal cord veins. They will be revealed by progressive or acute neurological symptoms.

We further separated SCAVMs into two types: (1) nidus type, in which an abnormal network is interspersed between arteries and veins; and (2) fistulas, in which a direct communication is seen between an artery and a vein. Although both types of lesions are found in the subpial space, niduses may be partially or totally buried in the spinal cord itself (Fig. 43.1). Conversely, large AVFs always remain superficial to the cord, as in the rest of the central nervous system, being located within the subarachnoid space, either dorsally or ventrally within the ventral sulcus in the subpial space. The venous drainage is likely to be mostly subpial in the former group, and subarachnoid in the latter.

In the pediatric age group, SCAVMs form the major part of the AVS. There are two types of the less often seen AVFs: the microAVFs, which have normalized draining veins, and the macroAVFs, which have enormously dilated draining veins. Greater than 80% of macroAVFs are associated with hereditary hemorrhagic telangiectasia (HHT), also called Rendu-Osler-Weber disease (Figs. 43.2, 3) (see Chapter 17).

Most spinal cord AVS in all age groups are single. However, 28% can be associated with some type of dysplasia, cutaneous vascular malformation, or spinal metameric (i.e., Cobb) syndrome (skin, vertebrae, and cord involvement at the same metameric level). In 9% of cases, HHT and a high-flow fistula were noted, and in 5% a Klippel-Trenaunay syndrome was found.

Radicular AVSs are rare. Some angioarchitectural appearances suggest a malformative AV sleeve around one nerve root, but in fact this corresponds to a congested radicular vein accompanying the spinal nerve.

43.2.1.2
Extradural Arteriovenous Shunts

These lesions are located in the epidural space. They are vascularized by dural or epidural branches of segmental arteries, and drain primarily into epidural venous plexuses. Neurological symptoms will occur if the venous drainage is rerouted towards spinal cord veins, thus creating a venous congestive myelopathy.

Fig. 43.1a–e. SCAVM in a 13-year-old female with left sided radicular pain. a Sagittal T2-weighted MR image shows flow voids of the nidus in the dorsal aspect of cord. b, c Transmedullary supply to dorsally located SCAVM. Presence of a network of draining veins emerging around the lower cervical nerves. d, e Ventral and contralateral dorsal pial arteries contribute to the dorsal SCAVM